CASE REPORTS

Pharyngeal Edema Associated with Arteritis

A Report of Two Cases

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ARTERITIS, of which there are many types, is not a common disease. Lesions may remain localized or may be manifestations of a generalized arteritis which may not become apparent for varying periods of time.[1] The signs and symptoms are varied, so that the condition may mimic many other processes. Recently two patients presented at this hospital with the main complaint of sore throat and later were found to have arteritis.

CASE 1.—E.M.W., a 72-year-old white man, was seen by the Otolaryngology Service in May 1967, with a history of a persistent sore throat, headaches, weight loss, general malaise and a mild chronic cough, as well as myalgia and arthralgia for several months. He was found to have pharyngeal edema, the uvula and parapharyngeal areas being markedly swollen. No purulent secretion or cervical lymphadenopathy was noted. Cultures were taken from the throat and only normal flora were identified. The erythrocyte sedimentation rate (ESR) was 60 mm. in one hour (Westergren). No muscular or joint abnormalities were found that could be considered pertinent. The only medication that the patient had been taking (and continues to take occasionally) was a compound of propoxyphene hydrochloride, acetylsalicylic acid, acetophenetidin and caffeine (Darvon Compound-65; Eli Lilly). There has never been any evidence that he was allergic to this drug. The patient later complained of a generalized pain on the left side of the forehead as well as a decrease in vision in the left eye. Funduscopic examination revealed a thrombosed central retinal artery; a central scotoma was present at that time and has persisted. No changes in the right eye were discovered then or since. The pharyngeal edema lasted for several weeks before a diagnosis of temporal arteritis was reached as the result of a biopsy from the left temporal artery.

The biopsy specimen showed a muscular artery surrounded by fibroadipose tissue. There was some change in the arterial wall, particularly in the media about the internal elastic lamina. Degenerative changes with fragmentation of the elastica were noted. No giant cells were seen. One section included a small vessel which was probably a branch of the temporal artery; this vessel, as well as showing degenerative changes in the media, was occluded by a thrombus. Although the histology did not present the classical picture of a giant-cell arteritis, the condition was a definite vasculitis and could be included in the group of temporal or cranial arteritis (Fig. 1).

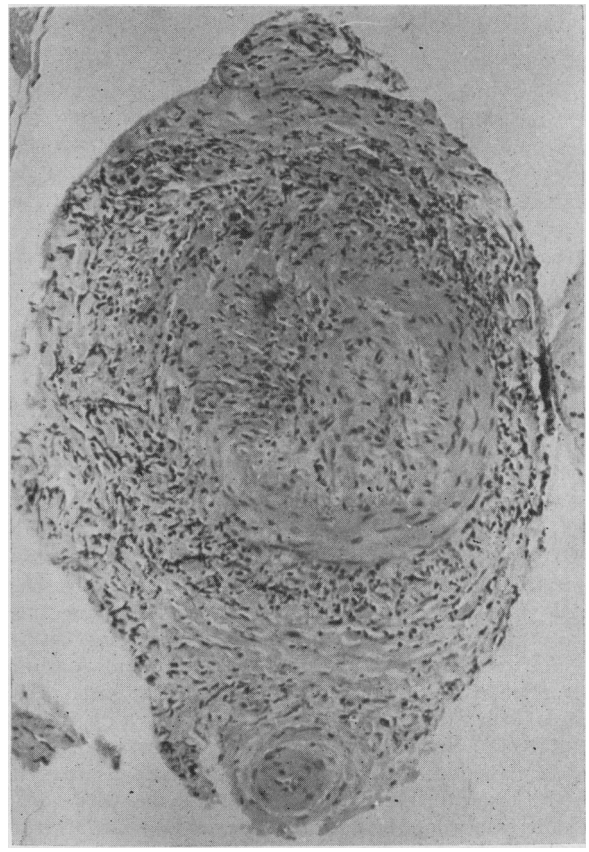

Fig. 1.—Cross-section of arteriole from Case 1 showing the typical vascular changes reported. (× 100.)

Once the diagnosis was established, the patient was given prednisone, 15 mg. four times daily. The dose was gradually decreased to the present maintenance dose of 5 mg. once a day. Occasionally the patient has a moderately severe attack of myalgia; he then increases the prednisone intake to 5 mg. twice daily and this is sufficient to control any long-lasting pain. The ESR fell after treatment was started.

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His sore throat, as well as his other symptoms, was immediately relieved once steroid therapy was initiated. At present (one and one-half years later) he has some mild intermittent myalgia and arthralgia as the only remaining manifestations of the condition. One year ago the patient had a mild myocardial infarction but this has imposed no residual restrictions on his activities; he is now back to his pre-infarct condition. This was the only cardiovascular change noted either before or after treatment. The patient's blood pressure and pulse rate were normal before and unchanged after the steroid therapy. He has not had a repeat attack of pharyngeal edema.

CASE 2.—C.P., a 64-year-old white man, first reported to this hospital in October 1964, with a history of weight loss (from 145 to 128 lbs. in a period of five months), intermittent edema in the legs, lethargy, left flank pain for approximately five years and a mild cough for two weeks. No diagnosis was made at that time. In March 1965 he was admitted with the same complaints. At this time, blood tests as well as bone and lymph node biopsies were carried out. The only abnormality reported was an ESR of 45 mm. in one hour (Westergren). He was discharged without any diagnosis.

On October 18, 1967, he was readmitted in acute distress with pharyngeal edema and gave a history of having had a "sore throat" for two weeks. He had not taken any medication and had no history of drug allergies. The uvula, hypopharynx and parapharyngeal areas were grossly edematous. He was unable to swallow owing to the swelling and pain. He was immediately given intravenous fluids which contained 100 mg. of hydrocortisone sodium succinate (Solu-Cortef, Upjohn) and antibiotics. His throat secretion yielded no abnormal flora when cultured. After several days of treatment his symptoms subsided dramatically and he was discharged from hospital. During his admission he was completely afebrile. He had no abnormal cardiovascular findings; his blood pressure and pulse were not increased.

Less than two months after he was discharged, a small tumour was removed from the left forearm. The report of the histological examination was as follows: The specimen consisted of adipose tissue with a small artery running through its centre. The walls of the artery and immediately adjacent tissues showed infiltration by acute inflammatory cells, with small numbers of eosinophils and chronic inflammatory cells. The lumen of the artery was thrombosed. The thrombosis appeared to be of different ages in different parts of the vessels. There was some evidence of luminal recanalization. The diagnosis was *periarteritis nodosa*. The picture was similar to that shown in the photomicrograph of Case 1.

DISCUSSION

Kussmaul and Maier[2] introduced the term "periarteritis nodosa" in 1866 to describe a systemic disease characterized by visible nodules along the course of medium-sized muscular arteries. The lesions were segmental in their distribution and to varying degrees involved most parts of the body. Thus the picture accounted for multiple clinical manifestations that seemed unrelated. Many theories of the cause of this vascular phenomenon have been proposed.[3-7] Allergic, infectious and toxic agents have all been suggested at one time or another as responsible. As yet no theory of causation has gained acceptance, but "hypersensitivity"—that is, an antigen-antibody reaction—seems to be favoured.[4, 6]

Zeek[8, 9] prefers the term "necrotizing angiitis" rather than polyarteritis, since she believes the condition comprises the following five subgroups: (1) periarteritis nodosa, (2) hypersensitivity angiitis, (3) rheumatic arteritis, (4) allergic granulomatous angiitis and (5) temporal or cranial arteritis.

The multiple symptoms that can be associated with this disease are illustrated by the following reported cases. In a group of 300 patients with polyarteritis nodosa[10] an analysis of the complaints showed that 85% presented with fever, 50% with coughs, 50% with generalized edema, 45% with weakness and/or weight loss, 30% with headaches and 10% with myalgia. Other recent reports show this same trend.[3, 11] All these symptoms and signs were seen to a greater or lesser extent in the cases reported here, except that no generalized edema was noted in either patient. The edema was entirely confined to the pharyngeal areas. I can offer no explanation why this should have been so.

In connection with Case 1 it should be noted that "cranial arteritis is part of a generalized vascular disorder characterized by granulomatous inflammation, but the involvement is so often confined to the arteries of the carotid system that its designation as a separate clinical entity is clearly indicated.

"Involvement of the temporal arteries is most frequent. Disease of the ophthalmic and retinal vessels is common, and the process may affect others, including the carotid, subclavian, coronary, renal, mesenteric and pulmonary arteries as well as those of the extremities.

"In about 20% of patients followed for several months the disease will assume an atypical form, and a variety of other manifestations may appear—the stage of systemic complications."[1]

The peak incidence of polyarteritis nodosa seems to occur in the fifth and sixth decades.[11] Twice as many males are affected as females. Steroids constitute the treatment of choice at present. Frohnert and Sheps[11] at the Mayo Cli-

nic reviewed 130 patients over a 16-year period and showed a 48% five-year survival rate for those treated with ACTH or corticosteroid therapy compared with only 13% for untreated patients. These results agree with those reported by the British Medical Research Council[12] but differ somewhat from the opinion expressed by Rupe.[13]

It is also interesting to note that in 4 of the 18 cases of necrotizing vasculitis reviewed by Owano and Sueper[3] there was involvement of veins as well as of small arteries and arterioles.

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Hodgkin's Disease Complicated by Infection with *Mycobacterium kansasii*

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PATIENTS with Hodgkin's disease have an increased incidence of tuberculosis. This was recognized by Ewing[1] in 1940: ". . . in New York, where the disease is very common, tuberculosis follows Hodgkin's disease like a shadow". In 1947 Jackson and Parker[2] cited a 20% incidence of active tuberculosis in patients with Hodgkin's disease. In 1959, Razis, Diamond and Craver[3] reported 20 cases of tuberculosis in a series of 1024 patients with Hodgkin's disease seen at The Memorial Hospital in New York, an incidence of 1.8%. Evidence of tuberculosis was found in 12 (5.1%) of 234 autopsies in that series.

The report below describes the first case of Hodgkin's disease complicated by pulmonary infection with *Mycobacterium kansasii.*

In November 1959, C.W., a 51-year-old white man, complained to his family physician of fatigue, weight loss and fever. Physical examination revealed enlarged axillary and anterior and posterior cervical lymph nodes. A chest radiograph showed hilar lymphadenopathy. In December 1959, Hodgkin's granuloma was diagnosed from a cervical node biopsy. The patient was treated with nitrogen mustard and a complete clinical remission of the disease followed. In July 1960, palpable nodes were found in the left cervical, left axillary and both inguinal regions. After radiotherapy there was another complete remission. In November 1960 and again in March 1962, enlargement of the lymph nodes recurred in the same areas but disappeared after additional treatment with nitrogen mustard. In February 1963, local radiotherapy was successful in the treatment of enlarged nodes in the right cervical and in both axillary areas.

In August 1963, the patient was referred to our hospital with symptoms of fever, night sweats, nausea and vomiting. Physical examination showed enlarged left posterior cervical nodes and left axillary nodes. The oral temperature was 103° F. The hemoglobin was 13.6 g. per 100 ml.; the leukocyte count was 2800 and the platelet count was 106,000 per c.mm. A lymphangiogram was normal. The patient was given cyclophosphamide 100 mg. daily. All symptoms and clinical evidence of disease disappeared. In June 1964 a spontaneous left pneumothorax developed; this was successfully managed by closed suction of the pleural cavity. His disease remained in complete remission while he was receiving cyclophosphamide (75 to 150 mg. daily, depending on the leukocyte count) until August 1964.

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